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A Predictive Study of Congenital Heart Disease and Need for Care

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For long-term planning in the delivery of health care, prevalence data are essential for budget estimates in terms both of distribution and training of manpower and fiscal responsibility. From incidence figures, from the knowledge of the natural history of congenital heart disease and from predicted population estimates it is possible to construct a model that reflects the prevalence of congenital heart disease. This has been done for the state of California; the methods used and the data gathered should prove useful nationally.

It is estimated that there were in 1975 in California 17,531 children under 21 years of age with congenital heart disease; 24 percent of these had ventricular septal defects and 23 percent had pulmonary stenosis, 11 percent had atrial septal defects and 9 percent had aortic stenosis; the other forms of congenital heart disease constituted the remaining 33 percent. Based on these estimates it is then possible to plan the medical resources necessary for optimal care.

ESTIMATES of the incidence of congenital cardiac malformations* are now available.[1-3] It is difficult, however, to use these estimates for the determination of the prevalence of congenital cardiac malformations† because the incidence data fail to include attrition through death or recovery. For long-term planning in the delivery of health care, prevalence data are essential for budget estimates in terms both of distribution of manpower and fiscal responsibility.

A recent report[4] to the California legislature on the California Crippled Children Services concludes that for determinations of case finding effectiveness and for the establishment of objectives and priorities for future program operations it is necessary to find a meaningful point prevalence of the handicapped population. The report goes on to say that it would be possible to obtain detailed and useful figures on the population by conducting an extensive state survey based on a health examination survey. The report, however, concludes that such a survey probably would be exorbitantly expensive in terms of its benefits.

The purpose of this study is to use incidence and attrition data to create a model that reflects the expected prevalence of congenital cardiac malformations in California. Requirements for the delivery and maintenance of health care can then be assessed.

Specifically the objects of this study are the following: (1) to assess the number of cases of congenital cardiovascular malformation for a given year in the California population under 21 years of age. (2) To delineate the different types

*Incidence is the occurrence of congenital heart disease during a given period.

†Prevalence represents the proportion of the population with congenital heart disease at a particular time. The general relationship of prevalence (P) and incidence (I) may be expressed as $P = I \times D$, where D is duration of the disease in an individual case expressed with same time units as I.

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TABLE 1.—*Predicted Population of California[7] and Estimated Live Births* for Selected Years Between 1975 and 1994*

	Population	Live Births
1975	21,206,000	329,000
1980	22,659,000	351,000
1985	24,363,000	378,000
1990	26,098,000	405,000
1995	27,726,000	429,753

*A live birth rate of 15.5 is assumed.

TABLE 2.—*Incidence of Congenital Heart Disease Based on a Mean Three-Year Observation of 54,765 Children[8]*

54,765	Live births
420	Children with congenital heart disease 7.7*
78	Children died before 28 days
50	Children died between 28 days and 1 year
20	Children died after 1 year
272	Children with congenital heart disease survived for an average of three years 5.3*

*Incidence of congenital heart disease per 1,000 live births

of congenital cardiac malformations assessed. (3) To show that such predictions may be used to assess the extent of required health care resources.

It is important to note that the population of California is almost a tenth of the total population of the United States. The United States Health Department's 1970 population figures for California and the nation were 20 million and 203 million people, respectively; the 1975 figures were 21 million and 213 million, respectively.

Materials and Methods

The population under consideration for this study is that of the state of California. From predictions of the number of live births in California from 1975 to 1994 it is possible by the use of incidence figures to estimate the number of children who would be born with congenital heart disease each year. Those children born in the final year and all the children from previous years who enter the second year of life are then added together over the 20-year period. The total estimate for the cases of congenital heart disease in 1994 is, however, much larger than the actual prevalence of congenital heart disease in the under 21-year-old children because this number fails to include either an estimated attrition through spontaneous improvement or death or to exclude those children who reach twenty years of age.

By the use of such a predictive study, which is the same length in years as the upper age limit of the studied cohort, one is able to ensure that at the end of the 20-year period, all the children included will be those who have been predicted and that all the children will be 20 years old or less.

A knowledge of the natural history and surgical management is then applied to the individual lesions. With respect to congenital heart disease the prevalence of heart disease in the 20-year-old and under group can therefore be regarded:

Prevalence of congenital heart disease in those under 21 years of age in California equals (yearly California population for 20 years × live birth rate : 1,000) × (incidence of congenital heart disease : 1,000) minus (the children expected to die in the first year of life in each of the first 19 years of the study + the spontaneous improvement rate + the premature death rate).

From California population predictions and live birth rates are determined the numbers of children expected to be born (Table 1). Incidence figures for congenital heart disease are then used to estimate the predicted number of cases (Table 2). For the years 1975 through 1994 all the children with congenital heart disease are added up.

From this total are subtracted all those born between 1975 and 1993 who fail to survive for one year, those who would be expected spontaneously to improve and those who would be expected prematurely to die. This figure would reflect the number of children up to the age of 20 with congenital heart disease in California in 1994. A prevalence ratio is then determined.

In order to avoid the necessity of carrying out an analysis for attrition through spontaneous improvement or death for each of the years between 1975 and 1994, a yearly estimate of survivors of children with congenital heart disease born in 1975 is then made (Table 3). Similarly, estimates are made for each individual cardiac lesion. These estimates are then proportionally increased by a population factor which represents the yearly increase in California population (Table 4). The total estimate of congenital heart disease can then be used to derive a prevalence ratio for 1994.

For children with ventricular septal defects and with pulmonary stenosis, the health care require-

TABLE 3.—*Yearly Estimate of Survivors of Children With Congenital Heart Disease Born in 1975*

Year	Year of Life	Number of Children	Population Factor*
1975	<1	2,520	1.29
1976	2	1,460	1.27
1977	3	1,304	1.25
1978	4	1,147	1.24
1979	5	1,104	1.23
1980	6	1,061	1.21
1981	7	1,018	1.19
1982	8	975	1.17
1983	9	933	1.16
1984	10	891	1.15
1985	11	849	1.13
1986	12	820	1.11
1987	13	791	1.10
1988	14	762	1.08
1989	15	733	1.07
1990	16	704	1.06
1991	17	675	1.04
1992	18	646	1.02
1993	19	617	1.01
1994	20	589	1.00

*Population factor is the population of California in 1994 divided by the population in the index year. For instance in 1980, the population factor $= \frac{27,435,400}{22,659,000} = 1.21$

TABLE 4.—*Estimated Numbers of Children With Congenital Heart Disease in California in 1995*

Year of Birth	Year of Life	Number of Children*
1994	1	3,251
1993	2	1,854
1992	3	1,630
1991	4	1,442
1990	5	1,358
1989	6	1,284
1988	7	1,211
1987	8	1,141
1986	9	1,082
1985	10	1,025
1984	11	959
1983	12	910
1982	13	870
1981	14	822
1980	15	784
1979	16	746
1978	17	702
1977	18	659
1976	19	623
1975	20	589
TOTAL		22,922

(Number of cases in 1995 × 100 ÷ California population in 1995) Prevalence Ratio = 0.078

*This number represents the childen who survived until 1995 from those born in the year listed alongside the number. The number of children is obtained in each year of birth by proportionately increasing the number of children in a certain year of life in Table 3 by the population factor. For instance, in Table 3 in the fifth year of life there are estimated to be 1,104 survivors of children born in 1975. However, children alive in 1995 who were in their fifth year of life would have been born in 1990. Consequently 1,104 × 1.23 (from Table 3) equals 1,358 (Table 4)

ments are estimated and compared with those actually rendered in California.

Major assumptions in this study include a relatively stable live birth rate, a predicted population growth, stable incidence figures for heart disease and a similar death rate in serious cardiac lesions and a constant prevalence ratio. A live birth rate of 15.5 per 1,000 population is used because it is the most recently available figure (1974)[5] and it is more likely to represent future figures rather than the high figures of the early 1970's.[6] The estimates for predicted population were obtained from the California State Department of Finance in Sacramento. The D-100 series was favored over other state and federal estimates because it takes into account not only the fertility rate but the migration rate and local factors.[7] The incidence data for the frequency for congenital heart disease were obtained from the Perinatal Research Branch, National Institutes of Health, Bethesda, Maryland.[3] The incidence of congenital heart disease in this study was estimated after data were obtained after a mean follow-up period of three years. In a further long-term study by Dr. Julien Hoffman, Professor of Pediatrics at the University of California, San Francisco, by 36 months of age over 98 percent of congenital heart disease had been located. It is realized, however, that the incidence figures for congenital heart disease are primarily obtained from a low socioeconomic group,[3] but the incidence figures are very similar to those obtained by Dr. Hoffman from the San Francisco Bay area of California. This latter group reflects all the major racial and socioeconomic groups with the exception of the very affluent.

The final assumptions were that the natural history studies of children with ventricular septal defects[8,9] reflected the distribution of small, moderate and large defects and that corrective cardiac surgical treatment results would not worsen.

It is to be emphasized that this study concerns itself with congenital heart disease and does not include rheumatic, atherosclerotic and hypertensive heart disease. Since accurate data are unavailable for the incidence of patent ductus arteriosus in premature children, bicuspid aortic valves and for prolapsed mitral valves, these three conditions have been omitted.

Results

Live births in California estimated from a live birth rate of 15.5 per thousand and from the predicted population are tabulated alongside repre-

sentative years (1975, 1980, 1985, 1990 and 1995 [Table 1]).

After the estimation of the live births per year is made, the incidence of specific cases of congenital heart disease is predicted. An example year is tabulated as Table 5. The lesions are listed under five columns. The first column is the total number of defects at birth, the next three columns represent, respectively, children who had cardiac lesions and died within 28 days of birth, between 28 days and one year, and after one year. The remaining column represents those children who would be expected to live longer than one year. Addition of these estimates indicates that there would be 40,038 children with congenital heart disease under 20 years of age in 1995.

This estimate, however, fails to include attrition through spontaneous improvement or death. Recovery of children with congenital heart disease is due predominantly to spontaneous closure of defects of the ventricular septum and closure of the ductus arteriosus. Mortality statistics for sudden death and from specific cardiac surgical procedures are also used.

As an example, Table 6 represents attrition in children born in 1975 with ventricular septal defects. By 1994 (when children born in 1975 are in their 20th year) it is expected that there would be at most 168 with ventricular septal defects. Each of the major groupings of cardiac conditions went through a similar analysis. It was estimated that each year there would be 411 children under the age of ten years who would undergo open heart surgery in California. This figure would include 80 children with pulmonary stenosis, 100 children with atrial septal defects, 60 children with coarctation of the aorta, 30 children with aortic stenosis, 66 children with the tetralogy of Fallot and 75 with miscellaneous conditions. It was estimated that 8 percent of these procedures would result in death. These figures are comparable to the figures prepared by the

TABLE 6.—*Estimated Attrition of Children Born in 1975 With Ventricular Septal Defects (VSD)*

1975	810 children born with VSD[3] ..	=810
1976	810 minus 84 children who die in first year[3] and 292 VSD that spontaneously close[8]	=434
1977	434 minus 170 VSD that close spontaneously which represent 75 percent of the remaining 227 small VSD's[6]	=264
1978-1984	264 minus 57 VSD's that close spontaneously which represents 25 percent of the remaining 227 small VSD's and 12 which represent deaths from attempted surgical closure 1984	=177
1985-1994	177 minus 9 which represent deaths from attempted surgical closure 1994	=168

TABLE 5.—*California Children Born With Congenital Heart Disease in 1975. Expected on Basis of Data Collected by Mitchell et al. 1973*[3]

	At Birth	*Died <28 Days*	*Died 28 Days to 1 Year*	*Died >1 Year*	*Survived for a Mean of Three Years*
Ventricular septal defect	810	54	30	18	708
Pulmonary stenosis	282	30	..	6	246
Atrial septal defect	186	42	..	..	144
Endocardial cushion defect	90	24	12	6	48
Patent ductus	210	.6	60	6	138
Coarctation of aorta	168	90	12	..	66
Tetralogy of Fallot	96	12	12	6	66
Aortic stenosis	96	..	..	..	96
Hypoplastic left heart	78	78	..	..	..
Endocardial fibroelastosis	60	6	18	24	12
Truncus arteriosus	42	36	..	..	6
Transposition of great arteries ...	66	18	30	6	12
Vascular ring	42	6	24	6	6
Single ventricle	18	..	12	..	6
Coronary abnormalities	30	12	12	6	..
Double outlet, right ventricle	24	..	6	12	6
Pulmonary atresia	30	6	24	..	..
Tricuspid atresia	30	12	12	..	6
Mitral regurgitation	18	..	..	6	12
Miscellaneous	144	36	36	18	54
TOTAL	2,520	468	300	120	1,632

Crippled Children Service Section of the State Department of Health in Sacramento for 1971, 1972 and 1973.[10,11] Each condition, however, was taken separately when it came to the mortality figures and a mean mortality figure for each condition was used in estimating operative mortality (see Table 7).

Most of the children who had ductus ligation would be lost to follow-up in the second decade and similarly 20 percent of children with surgically closed atrial septal defects. The rate of spontaneous closure of ductal ligation was likewise accounted for. This rate was 0.6 percent per annum after the age of 1 year.[12]

Table 8 lists in the first two columns the estimated incidence of congenital heart disease in live births for 1975.[3] Columns three through six list the estimated frequency of the eight groups of conditions for children born in 1975 at arbitrarily selected times. From these estimates it is possible to add total numbers of estimated cases of congenital heart disease in children born in 1975 who would be expected to live to 1994 and adding them to the total number of estimated cases of congenital heart disease in children born in 1976 who would live to 1994 (that is, in the 19th year). This procedure continued for all the year periods up to 1994. This gives the total number of expected congenital heart disease cases in 1994, namely 22,922.

In 1995 with a California population of more than 27 million a prevalence ratio can be estimated. Therefore with the assumption that the prevalence is relatively stable a total estimate for children with heart disease can be made for 1975 and a California population of just over 21 million (column seven, Table 8).

The maximum number of children with congenital heart disease who might be seen in 1975 is, therefore, 17,531. It is also possible to predict which types of lesions are in these children with congenital cardiovascular lesions (column seven, Table 8).

As an example of estimation of health care requirements the two commonest lesions have been selected, namely the ventricular septal defect and pulmonary stenosis. These two conditions together account for nearly half of the total number of estimated cases of congenital heart disease.

In 1975 there would be expected to be 810 children born with ventricular septal defects of which 36 percent[8] would probably be of at least moderate size (Table 9). On the basis of consultations with a pediatric cardiologist every two

TABLE 7.—*California Crippled Children Services Section Open Heart Surgery Figures* (Children 1-4, 5-20 years) From Which Mortality Estimates Were Derived (1971-1973)*[10,11]

		1971 (Percent)	*1972 (Percent)*	*1973 (Percent)*
59	Pulmonary stenosis	7	3	2
125	Atrial septal defect	1	1	1
41	Aortic stenosis	2	2	2
110	Tetralogy of Fallot	13	11	15
131	Ventricular septal defect ..	5	8	6
626	Total open heart cases ...	9	8	8

*These figures are derived from the 24 centers registered by the Crippled Children Services Program and do not reflect the mortality rates from the other 90 cardiac surgical centers in California.

TABLE 8.—*The Incidence, Frequency and Prevalence of Congenital Heart Disease in California*

The incidence is estimated for 1975, the frequency of congenital heart disease in 1976, 1978, 1984 and 1994 would be expected for those children born in 1975. The prevalence of congenital heart disease in 1975 is estimated.

	Incidence Among Live Births With Congenital Heart Disease in 1975		*Expected Frequency for Selected Years*				*Expected Prevalence 1975*	
	Number	*Percent*	*1976 (Percent)*	*1978 (Percent)*	*1984 (Percent)*	*1994 (Percent)*	*Number*	*Percent*
Ventricular septal defect	810	32	30	20	21	29	4,182	24
Pulmonary stenosis	282	11	17	21	28	30	4,003	23
Atrial septal defect	186	7	10	13	13.5	8	1,978	11
Aortic stenosis	96	4	6.5	8	11.5	16	1,637	9
Persistent ductus arteriosus ..	210	8	10	11	5	4	1,442	8
Coarctation of aorta	168	7	4.5	6	6	1	897	5
Tetralogy of Fallot	96	4	5	6	5	5	878	5
Miscellaneous	672	27	17	15	10	7	2,514	14
TOTAL NUMBERS*	2,520		1,460	1,147	849	587	17,531	

*Number of children with congenital heart disease who are expected to be born in 1975 and to survive to 1976, 1978, 1984 and 1994. The total number of predicted cases of congenital heart disease for 1975 is in the last column.

months for a year for this group and for the other 64 percent every four months, 3,300 visits might be anticipated. The children older than one year with ventricular septal defects are divided into five groups (after P. Wood)[9] and might be seen as follows:

Group I (a left to right shunt of 1.1 to 1.7:1) could be seen every two years and the remaining groups every six months. Group II (a left to right shunt between 1.8 and 2.9:1). Group III (a left to right shunt of over 3:1). Group IV (a left to right shunt between 1.5 and 3.5:1 and pulmonary hypertension) and group V (a left to right shunt of 0.5 to 1.5 and pulmonary hypertension).

In all, this would represent 7,770 patient visits to a cardiologist (Table 9).

From the group of children less than 1 year of age it is estimated that the majority of children with moderate shunts would be catheterized and about 20 would have a cardiac surgical procedure. From the over 1-year-old group (II and III) about 40 children per year would be expected to have corrective surgical operation and a similar number to have preoperative cardiac catheterizations.

Groups IV and V, children with a ventricular septal defect and pulmonary hypertension, have been prorated over the 20-year period and it is estimated that there would be about 25 catheterizations a year for each group and that the majority of group IV would have surgical operation (Table 9). Assuming a relatively stable surgery rate, there would be approximately 40 children in whom postoperative cardiac catheterizations would be required.

One patient visit each year for children with pulmonary stenosis and the assumption that 50 percent of the mild and all of the moderate and severe are eventually catheterized gives prorated estimates of 160 cardiac catheterizations per year and it is assumed that 50 percent of these children would have surgical operation (Table 9). In 1971, a total of 86 children had cardiac surgical operation on a stenotic pulmonary valve with approximately a 7 percent mortality.[10]

Discussion

There are both advantages and disadvantages in the use of the 20-year period 1975 through 1994 as opposed to the period 1956 through 1975 in the estimation of prevalence from incidence figures. Although by the use of this latter period, one can initiate the study with actual numbers of different types of congenital lesions, one is much less certain of attrition. Several major medical advances in the management of children were either perfected or first used between 1956 and 1975. Systemic to pulmonary anastomosis

TABLE 9.—*Estimated Health Care Requirements for Children in 1975 With Ventricular Septal Defect and Pulmonary Stenosis in California*

	Visits	*Catheterizations*	*Surgical Therapy*
Ventricular Septal Defect 4,182 cases			
Under 1 year[8]			
292 (36 percent) Spontaneous closure	900		
227 (28 percent) Became smaller	700	250	20
292 (36 percent) Moderate size	1,800		
Over 1 year[9]			
1,382 (41 percent) Group I L to R 1.1-1.7:1	700		
775 (23 percent) Group II L to R 1.8-2.9:1	1,500	80*	40
135 (4 percent) Group III L to R >3.0:1	270		
506 (15 percent) Group IV L to R 1.5-3.5:1 and pulmonary hypertension	1,000	25	25
573 (17 percent) Group V L to R 0.5-1.5:1 and pulmonary hypertension	1,000	25	
TOTAL 4,181	7,770	380	85
Pulmonary Stenosis 4,003 cases			
1,335 Mild			
1,334 Moderate			
1,334 Severe			
TOTAL 4,003	4,000	160†	80‡

*Includes 40 postoperative catheterizations per annum.
†On the basis of 50 percent of the mild pulmonary stenosis cases, all the moderate and severe will be catheterized over a 20-year period.
‡On the basis of 50 percent of the catheterization cases will have surgical operation.

was often lifesaving in situations of pulmonary oligemia. Open heart surgical operation to correct certain lesions became more practical. The mortality from transposition of the great arteries changed dramatically with the introduction of the balloon atrial septostomy (1966).

The importance of prevalence estimates of a disease process is evident when it is necessary to assess health care requirements. The health care requirements estimated in this study are probably overestimates as not all the cases will have been located and the attrition rate and surgical mortality may err on the conservative side.

The most recently available figures of the California Crippled Children Services pertaining to congenital heart disease were then compared with the estimates.

In the annual report of cardiac centers (1971 through 1973)[10,11] it is indicated that there were an average of 131 cardiac surgical closures of ventricular septal defects each year with a mortality of 6 percent in children under 21 years of age, figures similar to those estimated.

This report also indicates that just over 900 cardiac catheterizations were carried out in children under one year and 1,854 in children between 1 and 20 years of age. Although specific lesions are not commented upon, these figures would be compatible with the above estimates.

The assumptions made in this study probably err on the side of overestimating the population of specific congenital heart disease described in this study. It is to be reemphasized that the present estimates of the frequency of three conditions —persistent ductus arteriosus, bicuspid aortic valve and prolapsed mitral valve—are not accurate enough to be included in this study. However, this may in part be balanced by the genetic implications of increased survival of a group of patients with congenital heart disease.

It should also be noted that it is expected that the majority of children who have had cardiac surgical procedures will need to be studied further because of the high frequency of residua.[13,14]

Implications

Disease prevalence data are essential for the planning of health care delivery. The manpower and fiscal responsibilities can only be determined by a knowledge of the prevalence of disease process and the future predictions of the disease.

This paper describes an original method for determining prevalence estimates from incidence data. This method may be applied to any disease process which has a well-known natural history. Specifically, this paper predicts the amount of congenital heart disease in California for the next 20 years. It is important to realize that merely by increasing the estimates tenfold, the study is applicable to the whole of the United States.

Summary

In conclusion, this study has estimated the prevalence of congenital heart disease in California. To convert these estimates to apply to the whole of the United States of America, a population factor of ten times is required.

The study has outlined probable health care requirements for the two most common cardiac conditions which represent nearly 50 percent of the congenital cardiac lesions. Finally, this study may serve as a model on which prevalence estimates can be determined for future health care delivery.

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